# Chapter 33

# Assessment and Mangement of patients with Hematologic disorders

# Hematologic System

**Structures and Functions**

* **Hematology** is the study of blood and blood-forming tissues. This includes the bone marrow, blood, spleen, and lymph system.
* Blood cell production (**hematopoiesis**) occurs within the bone marrow. **Bone marrow** is the soft material that fills the central core of bones.
* Blood is a type of connective tissue that performs three major functions: transportation, regulation, and protection. There are two major components to blood: plasma and blood cells.
* Plasma is composed primarily of water, but it also contains proteins, electrolytes, gases, nutrients, and waste.
* There are three types of blood cells: erythrocytes (RBCs), leukocytes (WBCs), and thrombocytes (platelets).
* **Erythrocytes** are primarily composed of a large molecule called hemoglobin. **Hemoglobin,** a complex protein-iron compound composed of heme (an iron compound) and globin (a simple protein), functions to bind with oxygen and carbon dioxide.
* **Leukocytes** (WBCs) appear white when separated from blood. There are five different types of leukocytes, each of which has a different function.
  + Granulocytes (neutrophils, eosinophils, basophils): the primary function of the granulocytes is **phagocytosis,** a process by which WBCs ingest or engulf any unwanted organism and then digest and kill it. The neutrophil is the most common type of granulocyte.
  + Lymphocytes: the main function of lymphocytes is related to the immune response. Lymphocytes form the basis of the cellular and humoral immune responses.
  + Monocytes: monocytes are phagocytic cells. They can ingest small or large masses of matter, such as bacteria, dead cells, tissue debris, and old or defective RBCs.
  + Leukocytosis- ^ lvl of Leukocytes
  + Leukocytopenia- decreased lvls of Leukocytes in circulation
* The primary function of **thrombocytes,** or platelets*,* is to initiate the clotting process by producing an initial platelet plug in the early phases of the clotting process.
* **Hemostasis** is a term used to describe the blood clotting process. This process is important in minimizing blood loss when various body structures are injured.
* Four components contribute to normal hemostasis: vascular response, platelet plug formation, the development of the fibrin clot on the platelet plug by plasma clotting factors, and the ultimate lysis of the clot.
* Another component of the hematologic system is the spleen, which is located in the upper left quadrant of the abdomen. The functions of the spleen can be classified into four major functions: hematopoietic, filtration, immunologic, and storage.
* The lymph system—consisting of lymph fluid, lymphatic capillaries, ducts, and lymph nodes—carries fluid from the interstitial spaces to the blood.

**Assessment**

* Much of the evaluation of the hematologic system is based on a thorough health history, and a number of health patterns should be assessed.
* A complete physical examination is necessary to accurately examine all systems that affect or are affected by the hematologic system, including an assessment of lymph nodes, liver, spleen, and skin.

**Diagnostic Studies**

* The most direct means of evaluating the hematologic system is through laboratory analysis and other diagnostic studies.
* The complete blood count (CBC) involves several laboratory tests, each of which serves to assess the three major blood cells formed in the bone marrow.
* Erythrocyte sedimentation rate (ESR or “sed rate”) measures the sedimentation or settling of RBCs and is used as a nonspecific measure of many diseases, especially inflammatory conditions.
* The laboratory tests used in evaluating iron metabolism include serum iron, total iron-binding capacity (TIBC), serum ferritin, and transferrin saturation.
* Radiologic studies for the hematology system involve primarily the use of computed tomography (CT) or magnetic resonance imaging (MRI) for evaluating the spleen, liver, and lymph nodes.
* Bone marrow examination is important in the evaluation of many hematologic disorders. The examination of the marrow may involve aspiration only or aspiration with biopsy.
* Lymph node biopsy involves obtaining lymph tissue for histologic examination to determine the diagnosis, and to help for planning therapy.
* Testing for specific genetic or chromosomal variations in hematologic conditions is often helpful in assisting in diagnosis and staging. These results also help to determine the treatment options and prognosis.

Nursing Management: Hematologic Problems

**ANEMIA**

* **Anemia** is a deficiency in the number of erythrocytes (red blood cells [RBCs]), the quantity of hemoglobin, and/or the volume of packed RBCs (hematocrit), which can lead to tissue hypoxia.
* Hemoglobin (Hb) levels are often used to determine the severity of anemia.
* Correcting the cause of the anemia is ultimately the goal of therapy.
* Interventions may include blood or blood product transfusions, drug therapy, volume replacement, oxygen therapy, dietary modifications, and lifestyle changes.

**Anemia Caused By Decreased Erythrocyte Production**

***Iron-Deficiency Anemia***

* **Iron-deficiency anemia** may develop from inadequate dietary intake, malabsorption, blood loss, or hemolysis. Also, pregnancy contributes to iron deficiency because of the diversion of iron to the fetus for erythropoiesis, blood loss at delivery, and lactation.
* The main goal of collaborative care for iron-deficiency anemia is to treat the underlying disease causing reduced intake (e.g., malnutrition, alcoholism) or absorption of iron. In addition, efforts are directed toward replacing iron with dietary changes or supplementation.
* It is important for a nurse to recognize groups of individuals who are at an increased risk for the development of iron-deficiency anemia. These include premenopausal and pregnant women, persons from lower–class socioeconomic backgrounds, older adults, and individuals experiencing blood loss.

***Thalassemia***

* **Thalassemia** is a group of diseases that has an autosomal-recessive genetic basis that involves inadequate production of normal hemoglobin.
* An individual with thalassemia may have a heterozygous or homozygous form of the disease, based on the number of thalassemia genes the individual has.
* Thalassemia minor requires no treatment because the body adapts to the reduced level of normal hemoglobin.
* The symptoms of thalassemia major are managed with blood transfusions or exchange transfusions in conjunction with IV deferoxamineto reduce the iron overloading (**hemochromatosis**) that occurs with chronic transfusion therapy.

# Megaloblastic Anemias

* **Megaloblastic anemias** are a group of disorders caused by impaired DNA synthesis and characterized by the presence of large RBCs.
* Macrocytic (large) RBCs are easily destroyed because they have fragile cell membranes.
* Two common forms of megaloblastic anemia are cobalamin deficiency and folic acid deficiency.
  + Cobalamin (vitamin B12) deficiency is most commonly caused by **pernicious anemia,** which results in poor cobalamin absorption through the GI tract. Parenteral or intranasal administration of cobalamin is the treatment of choice.
  + Folic acid (folate) is required for DNA synthesis leading to RBC formation and maturation and therefore can lead to megaloblastic anemia. Folic acid deficiency is treated by replacement therapy.

***Aplastic Anemia***

* **Aplastic anemia** is a disease in which the patient has peripheral blood *pancytopenia* (decrease of all blood cell types) and hypocellular bone marrow.
* Management of aplastic anemia is based on identifying and removing the causative agent (when possible) and providing supportive care until the pancytopenia reverses.

**Anemia Caused By Blood Loss**

***Acute Blood Loss***

* Acute blood lossoccurs as a result of sudden hemorrhage.
* Causes of acute blood loss include trauma, complications of surgery, and conditions or diseases that disrupt vascular integrity.
* Collaborative care is initially concerned with replacing blood volume to prevent shock and identifying the source of the hemorrhage and stopping the blood loss.

***Chronic Blood Loss***

* The sources of chronic blood loss are similar to those of iron-deficiency anemia (e.g., bleeding ulcer, hemorrhoids, menstrual and postmenopausal blood loss).
* Management of chronic blood loss anemia involves identifying the source and stopping the bleeding. Supplemental iron may be required.

**Anemia Caused By Increased Erythrocyte Destruction (Hemolytic Anemia)**

# Sickle Cell Disease

* **Sickle cell disease** is a group of inherited, autosomal recessive disorders characterized by the presence of an abnormal form of hemoglobin in the erythrocyte.
* The major pathophysiologic event of this disease is the sickling of RBCs. Sickling episodes are most commonly triggered by low oxygen tension in the blood.
* With repeated episodes of sickling there is gradual involvement of all body systems, especially the spleen, lungs, kidneys, and brain.
* Collaborative care for a patient with sickle cell disease is directed toward alleviating the symptoms from the complications of the disease and minimizing end target-organ damage. There is no specific treatment for the disease.

***Acquired Hemolytic Anemia***

* Extrinsic causes of hemolysis can be separated into three categories: (1) physical factors, (2) immune reactions, and (3) infectious agents and toxins.
* Physical destruction of RBCs results from the exertion of extreme force on the cells.
* Antibodies may destroy RBCs by the mechanisms involved in antigen-antibody reactions.
* Infectious agents foster hemolysis in four ways: (1) by invading the RBC and destroying its contents, (2) by releasing hemolytic substances, (3) by generating an antigen-antibody reaction, and (4) by contributing to splenomegaly as a means of increasing removal of damaged RBCs from the circulation.

**Hemochromatosis**

* **Hemochromatosis** is an autosomal recessive disease characterized by increased intestinal iron absorption and, as a result, increased tissue iron deposition.
* The goal of treatment is to remove excess iron from the body and minimize any symptoms the patient may have.

**Polycythemia**

* **Polycythemia** is the production and presence of increased numbers of RBCs. The increase in RBCs can be so great that blood circulation is impaired as a result of the increased blood viscosity and volume.
* Treatment is directed toward reducing blood volume/viscosity and bone marrow activity. Phlebotomy is the mainstay of treatment.

**THROMBOCYTOPENIA**

* **Thrombocytopenia** is a reduction of platelets below 150,000/μl (150 × 109/L).
* Platelet disorders can be inherited, but the vast majority of them are acquired. The causes of acquired disorders include autoimmune diseases, increased platelet consumption, splenomegaly, marrow suppression, and bone marrow failure.
* The most common acquired thrombocytopenia is a syndrome of abnormal destruction of circulating platelets termed *immune thrombocytopenic purpura* (ITP). Multiple therapies are used to manage the patient with ITP, such as corticosteroids or splenectomy.
* One of the risks associated with the broad and increasing use of heparin is the development of the life-threatening condition called *heparin-induced thrombocytopenia and thrombosis syndrome* (HITTS). Heparin must be discontinued when HITTS is first recognized, which is usually if the patient’s platelet count has fallen 50% or more from its baseline or if a thrombus forms while the patient is on heparin therapy.
* For the nurse, the overall goals are that the patient with thrombocytopenia will (1) have no gross or occult bleeding, (2) maintain vascular integrity, and (3) manage home care to prevent any complications related to an increased risk for bleeding.

## HEMOPHILIA AND VON WILLEBRAND DISEASE

* **Hemophilia** is a sex-linked (men) recessive genetic disorder caused by defective or deficient coagulation factor. The two major forms of hemophilia, which can occur in mild to severe forms, are hemophilia A and hemophilia B.
* Von Willebrand disease is a related disorder involving a deficiency of the von Willebrand coagulation protein.
* Replacement of deficient clotting factors is the primary means of supporting a patient with hemophilia. In addition to treating acute crises, replacement therapy may be given before surgery and before dental care as a prophylactic measure.
* Home management is a primary consideration for the patient with hemophilia because the disease follows a progressive, chronic course.
* The patient with hemophilia must be taught to recognize disease-related problems and to learn which problems can be resolved at home and which require hospitalization.

**DISSEMINATED INTRAVASCULAR COAGULATION**

* **Disseminated intravascular coagulation** (DIC) is a serious bleeding and thrombotic disorder.
* It results from abnormally initiated and accelerated clotting. Subsequent decreases in clotting factors and platelets ensue, which may lead to uncontrollable hemorrhage.
* DIC is always caused by an underlying disease or condition. The underlying problem must be treated for the DIC to resolve.
* It is important to diagnose DIC quickly, stabilize the patient if needed (e.g., oxygenation, volume replacement), institute therapy that will resolve the underlying causative disease or problem, and provide supportive care for the manifestations resulting from the pathology of DIC itself.

**NEUTROPENIA**

* **Neutropenia** is a reduction in neutrophils, a type of granulocyte, and therefore is sometime referred to as granulocytopenia. The neutrophilic granulocytes are closely monitored in clinical practice as an indicator of a patient’s risk for infection.
* Neutropenia is a clinical consequence that occurs with a variety of conditions or diseases. It can also be an expected effect, a side effect, or an unintentional effect of taking certain drugs.
* Occasionally the cause of the neutropenia can be easily treated (e.g., nutritional deficiencies). However, neutropenia can also be a side effect that must be tolerated as a necessary step in therapy (e.g., chemotherapy, radiation therapy). In some situations the neutropenia resolves when the primary disease is treated (e.g., tuberculosis).
* The nurse needs to monitor the neutropenic patient for signs and symptoms of infection and early septic shock.

**MYELODYSPLASTIC SYNDROME**

* **Myelodysplastic syndrome** (MDS) is a group of related hematologic disorders characterized by a change in the quantity and quality of bone marrow elements. Although it can occur in all age groups, the highest prevalence is in people over 60 years of age.
* Supportive treatment consists of hematologic monitoring, antibiotic therapy, or transfusions with blood products. The overall goal is to improve hematopoiesis and ensure age-related quality of life.

**LEUKEMIA**

* **Leukemia** is the general term used to describe a group of malignant disorders affecting the blood and blood-forming tissues of the bone marrow, lymph system, and spleen.

* Classification of leukemia can be done based on acute versus chronic and on the type of WBC involved, whether it is of myelogenous origin or of lymphocytic origin.
  + The onset of acute myelogenous leukemia (AML) is often abrupt and dramatic. AML is characterized by uncontrolled proliferation of myeloblast, the precursors of granulocytes.
  + Acute lymphocytic leukemia (ALL) is the most common type of leukemia in children.
  + Chronic myelogenous leukemia(CML) is caused by excessive development of mature neoplastic granulocytes in the bone marrow, which move into the peripheral blood in massive numbers and ultimately infiltrate the liver and spleen. The natural history of CML is a chronic stable phase, followed by the development of a more acute, aggressive phase referred to as the blastic phase*.*
  + Chronic lymphocytic leukemia (CLL) is characterized by the production and accumulation of functionally inactive but long-lived, small, mature-appearing lymphocytes. The lymphocytes infiltrate the bone marrow, spleen, and liver, and lymph node enlargement is present throughout the body.
  + Hairy cell leukemia is a chronic disease of lymphoproliferation predominantly involving B lymphocytes that infiltrate the bone marrow and spleen. Cells have a “hairy” appearance under the microscope.
* Once a diagnosis of leukemia has been made, collaborative care is focused on the initial goal of attaining remission. In some cases, such as nonsymptomatic patients with CLL, watchful waiting with active supportive care may be appropriate.
* Cytotoxic chemotherapy is the mainstay of the treatment for leukemia. Hematopoietic stem cell transplantation is another type of therapy used for patients with different forms of leukemia.
* The overall nursing goals are that the patient with leukemia will (1) understand and cooperate with the treatment plan, (2) experience minimal side effects and complications associated with both the disease and its treatment, and (3) feel hopeful and supported during the periods of treatment, relapse, or remission.

**LYMPHOMAS**

* **Lymphomas** are malignant neoplasms originating in the bone marrow and lymphatic structures resulting in the proliferation of lymphocytes.
* There are two major types of lymphomas—Hodgkin’s lymphoma and non-Hodgkin’s lymphoma (NHL).

**Hodgkin’s Lymphoma**

* **Hodgkin’s lymphoma,** also calledHodgkin’s disease**,** is a malignant condition characterized by proliferation of abnormal giant, multinucleated cells, called *Reed-Sternberg cells,* which are located in lymph nodes.
* Although the cause of Hodgkin’s lymphoma remains unknown, the main interacting factors include infection with Epstein-Barr virus, genetic predisposition, and exposure to occupational toxins. The incidence of Hodgkin’s lymphoma is increased in incidence among human immunodeficiency virus infected patients.
* The nursing care for Hodgkin’s lymphoma is largely based on managing problems related to the disease (e.g., pain due to tumor), pancytopenia, and other side effects of therapy.

**Non-Hodgkin’s Lymphomas**

* **Non-Hodgkin’s lymphomas** (NHLs) are a heterogeneous group of malignant neoplasms of primarily B or T cell origin affecting all ages. A variety of clinical presentations and courses are recognized from indolent (slowly developing) to rapidly progressive disease.
* NHLs can originate outside the lymph nodes, the method of spread can be unpredictable, and the majority of patients have widely disseminated disease at the time of diagnosis.
* Treatment for NHL involves chemotherapy and sometimes radiation therapy. Nursing care is largely based on managing problems related to the disease (e.g., pain due to the tumor, spinal cord compression, tumor lysis syndrome), pancytopenia, and other side effects of therapy.

## MULTIPLE MYELOMA

* **Multiple myeloma,** or *plasma cell myeloma,* is a condition in which neoplastic plasma cells infiltrate the bone marrow and destroy bone.
* Multiple myeloma develops slowly and insidiously. The patient often does not manifest symptoms until the disease is advanced.
* Multiple myeloma is seldom cured, but treatment can relieve symptoms, produce remission, and prolong life. Chemotherapy is usually the first treatment recommended for multiple myeloma.
* Maintaining adequate hydration is a primary nursing consideration to minimize problems from hypercalcemia. Because of the potential for pathologic fractures, the nurse must be careful when moving and ambulating the patient.

**BLOOD COMPONENT THERAPY**

* Blood component therapy is frequently used in managing hematologic diseases. However, blood component therapy only temporarily supports the patient until the underlying problem is resolved.
* When the blood or blood components have been obtained from the blood bank, positive identification of the donor blood and recipient must be made. Improper product-to-patient identification causes 90% of hemolytic transfusion reactions.
* The blood should be administered as soon as it is brought to the patient. It should not be refrigerated on the nursing unit.
* Autotransfusion*,* or autologous transfusion, consists of removing whole blood from a person and transfusing that blood back into the same person. The problems of incompatibility, allergic reactions, and transmission of disease can be avoided.
* A blood transfusion reactionis an adverse reaction to blood transfusion therapy that can range in severity from mild symptoms to a life-threatening condition. Blood transfusion reactions can be classified as acute or delayed.

Acute Transfusion Reactions

* **Stop 1st , take VS then notify Doctor, maintain IV line w/ normal saline**
* **Unused blood must go back to bank b4 4hrs. (According to hospital policy)**
* The most common cause of hemolytic reactions is transfusion of ABO-incompatible blood.
* Febrile reactions are most commonly caused by leukocyte incompatibility. Many individuals who receive five or more transfusions develop circulating antibodies to the small amount of WBCs in the blood product.

The diagnosis of a febrile nonhemolytic reaction is made

by excluding other potential causes, such as a hemolytic reaction

or bacterial contamination of the blood product.

The signs and symptoms of a febrile nonhemolytic transfusion

reaction are chills (minimal to severe) followed by

fever (more than 1\_C elevation). The fever typically begins

within 2 hours after the transfusion is begun. Although the

reaction is not life-threatening, the fever, and particularly

the chills and muscle stiffness, can be frightening to the

* patient.
* Allergic reactions result from the recipient’s sensitivity to plasma proteins of the donor’s blood. These reactions are more common in an individual with a history of allergies.
* An individual with cardiac or renal insufficiency is at risk for developing circulatory overload. This is especially true if a large quantity of blood is infused in a short period of time, particularly in an elderly patient.
* Transfusion-related lung injuryis characterized by the sudden development of noncarcinogenic pulmonary edema (acute lung injury).
* An acute complication of transfusing large volumes of blood products is termed massive blood transfusion reaction*.* Massive blood transfusion reactions can occur when replacement of RBCs or blood exceeds the total blood volume within 24 hours.

**Delayed Transfusion Reactions**

* Delayed transfusion reactions include delayed hemolytic reactions, infections, iron overload, and graft-versus-host disease.
* Infectious agents transmitted by blood transfusion include hepatitis B and C viruses, HIV, human herpesvirus type 6, Epstein-Barr virus, human T cell leukemia, cytomegalovirus, and malaria.
* 6hrs to 14 days – must report 2 RN @ change of shift to continue monitoring.